An Aberrant Subclavian Artery Exhibiting the Partial Steal Phenomenon in a Patient with VACTERL Association

Hrvoje Budincevic 1, Katarina Starcevic 2, Ivan Bielen 3 and Vida Demarin 4

Abstract

We herein report the case of a 22-year-old Caucasian man with known vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, cardiac defects, renal and limb anomalies (VACTERL) association who presented with a headache and vertigo following the sudden and temporary loss of consciousness while attending a concert four days before admission to the hospital. On a physical examination, the following findings were found: a low body height, low-set ears, thoracic scoliosis and a mild holosystolic heart murmur. A neurosonological examination revealed a partial subclavian steal phenomenon. CT angiography of the neck vessels and aortic arch confirmed an anomalous right subclavian artery—known as the lusorian artery. Further studies are warranted in patients with VACTERL in order to identify possible links between the prevalence of an aberrant right subclavian artery (lusorian artery) and possible congenital subclavian steal syndrome or dysphagia lusoria. In addition, duplex ultrasound of the carotid and vertebral arteries may be performed as part of screening examinations in patients with congenital syndromes.

Key words: VACTERL, subclavian steal syndrome, lusorian artery

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Introduction

Subclavian steal syndrome refers to a pathologic condition in which, as a result of proximal subclavian artery stenosis or occlusion, there is a retrograde flow through the vertebral artery which thus causes insufficient brain perfusion and subsequent transient neurologic symptoms due to cerebral ischemia (1).

Vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, cardiac defects, renal and limb anomalies (VACTERL) is an association diagnosed when at least three of the following anomalies are present in a newborn: vertebral defects, anal atresia, cardiovascular anomalies (ventricular septal defect being the most common), esophageal atresia, renal anomalies and limb defects (2).

The aim of this case report is to present a patient with a congenital syndrome known as VACTERL association with an aberrant subclavian artery and consequent partial subclavian steal phenomenon.

Case Report

We herein report the case of a 22-year-old Caucasian man who presented with a headache and vertigo following the sudden and temporary loss of consciousness while attending a concert four days before admission to the hospital. His prior medical history included surgical repair of esophageal atresia as a newborn, with a verified ventricular septal defect on heart ultrasound.

The findings of a complete physical exam were unremarkable, with the exception of a low body height, low-set ears, thoracic scoliosis and a mild holosystolic heart murmur. The patient’s blood pressure was normal, without differences in the arm blood pressures or symptoms of arm claudication. A chest X-ray detected significant scoliosis at the cervicothoracic junction with prominent rib overlap at the C5-C6 level. Meanwhile, brain nuclear magnetic resonance (NMR) was unremarkable; however, NMR of the cervical spine showed the following vertebral anomalies: vertebral block between...
Figure 1. a: Duplex ultrasound of the right vertebral artery showing a reduced blood flow velocity through the right vertebral artery, with signs of a mid-systolic cleft corresponding to a type 3 waveform (arrows). b: Blood pressure cuff maneuver duplex ultrasound of the right vertebral artery showing a reduced blood flow velocity through the right vertebral artery, with signs of a mid-systolic cleft with mild retrograde flow following deflation of the cuff on the right arm (arrows). c: Transcranial duplex ultrasound of the right vertebral artery (arrow) showing a mid-systolic notch with a type 2 waveform (arrow).

Discussion

The presence of congenital partial subclavian steal syndrome has not yet been reported in patients with VACTERL association. When diagnosing subclavian steal phenomenon, duplex ultrasound of the carotid and vertebral arteries enables clinicians to recognize and differentiate between the pre-steal, partial steal and complete steal phenomena (4). The type of waveform on duplex ultrasound of the vertebral arteries corresponds to the severity of the subclavian steal phenomenon (3). The most common cause of subclavian steal syndrome is atherosclerosis (5); other unusual causes include arteriopathy (Takayasu’s disease, temporal arteritis) and congenital lesions of the aortic arch or subclavian artery (5, 6). Symptoms resulting from subclavian steal syndrome generally include arm claudication, paroxysmal vertigo, syncope, drop attacks, dizziness, diplopia, ataxia and dysarthria (5).

The presence of a lusorian artery is one of the most common aortic arch anomalies, with an incidence of 0.5-2.6% (7). It consists of a right subclavian artery originating as the most distal aortic arch branch, most commonly with a retroesophageal course, thus sometimes causing dysphagia difficulties (known as dysphagia lusoria) due to compression of the esophagus (8).

The partial form of subclavian steal syndrome has been described in the literature only once, whereas the occurrence of complete subclavian steal syndrome resulting from an aberrant lusorian artery has been presented several times (8-11).

Dysphagia is a common symptom in patients with VACTERL association; however, most patients with dysphagia have been previously treated for tracheoesophageal fistula with surgery (12). Further studies are warranted in patients with VACTERL association in order to identify any possible links between this syndrome, including the presence of a lusorian artery, and the incidence of symptomatic dysphagia lusoria and eventually the subclavian steal phenomenon.

In the present case, no structural stenosis of the right subclavian or vertebral artery was found on CTA. Digital subtraction angiography (DSA) was not performed because the right vertebral artery, with a mid-systolic cleft corresponding to the partial subclavian steal phenomenon with a type 3 waveform in the extracranial segment of the vertebral artery (3). The blood pressure cuff maneuver was performed on the right arm, and, following deflation of the cuff, the mid-systolic cleft became more pronounced with a minimal retrograde flow during systole (Fig. 1b). Transcranial color coded ultrasound showed a mid-systolic notch corresponding to a type 2 waveform (Fig. 1c). Computed tomography angiography (CTA) of the neck vessels and aortic arch confirmed an anomalous right subclavian artery originating as the most distal branch of the aortic arch and coursing in a retroesophageal fashion (lusorian artery), with mild compression of the esophagus (Fig. 2).
patient’s symptoms resolved during hospitalization and due to the risks of invasive angiography.

The subclavian steal phenomenon, as observed in this case, was likely due to the anatomical position of the aberrant right subclavian artery and site of origination of the right vertebral artery (Fig. 2a), which was further compromised with compression of the esophagus and the course of the right vertebral artery (Fig. 2d) through the scoliotic cervical spine due to the patient’s vertebral anomaly. The patient’s cardiac ventricular septal defect also likely played a role in this case, as this condition can be found in patients with congenital subclavian steal syndrome (6).

Duplex ultrasound of the carotid and vertebral arteries is a useful screening tool in patients with congenital syndromes.

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References